An Atypical Case of an Ulcerative Lesion of the Naris and Nasal Septum
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Abstract

Educational Objective: At the conclusion of this presentation, the participants should be able to discuss the differential diagnosis for an erosive cutaneous facial lesion and devise a diagnostic approach to these lesions. Objectives: To present a rare case of an ulcerative nasolabial cutaneous lesion eroding through the anterior nasal septum, and discuss the differential diagnosis for this atypical lesion. Study Design: Case report and literature review. Methods: A 62 year old woman referred for evaluation of an incidental thyroid nodule was found to have an unrelated, large, ulcerative mass involving her upper right lip and extending into the nasal vestibule with erosion of the cartilaginous nasal septum. We suspected a malignant cutaneous neoplasm and performed a punch biopsy of this lesion. Over the following 10 months two repeat biopsies were performed, and all three revealed acute and chronic inflammation without evidence of malignancy. Results: Over an 11 month period a multidisciplinary effort involving otolaryngology, pathology, radiology, rheumatology, and infectious disease experts at our tertiary referral center have been unable to make a definitive diagnosis for this patient’s lesion. Microbiology and pathology studies have been non-definitive. A mildly positive ANCA level prompted consideration of Wegener’s granulomatosis, however this was deemed unlikely given an unusual immunological staining pattern, absence of vasculitis, and complete lack of other disease symptoms. The patient has been empirically treated with high dose corticosteroids, which have resulted in dramatic and ongoing improvements. Our presentation will include radiological imaging and photographic documentation of her lesion’s progression. Conclusions: While erosive lesions of the head and neck immediately raise concern for cutaneous malignancy, the differential diagnosis should remain broad, and a multidisciplinary approach is useful in targeting diagnostic testing and treatment.

Introduction

Destructive midline facial lesions are a difficult diagnostic dilemma based on a wide differential diagnosis with drastically varying treatments dependant on etiology.

• First case series of these lesions reported by Stewart in 1922.
• Lab tests, biopsies with histological analysis (including fresh specimens), and tissue for infectious workup are the mainstays of a thorough diagnostic workup.
• There are a proportion of patients with an unknown etiology for these lesions even after extensive, multidisciplinary workup, known as idiopathic midline destructive diseases.
• Treatment includes resection, antimicrobials, immunomodulators, chemo or radiation therapy, or cessation of offending toxic agents (cocaïne) – based on etiology.
• In idiopathic cases empiric treatment should begin with less invasive/toxic treatments and progress to more invasive therapies for lesions that are refractory to initial therapy.

Case Presentation

HPI: 62-year-old caucasian female presented for evaluation of an incidental thyroid nodule, and found to have an unrelated ulcerative lesion of right nasal aperture extending to the upper lip. Per patient the lesion was present since nasal trauma with airbag impact during motor vehicle accident four months prior. Patient with history of minimal remote tobacco use, denied cocaine use, no personal history of cutaneous malignancies or radiation exposure, no family history of malignancy or autoimmune disease, no kidney disease or pulmonary complaints.

Physical Exam: Revealed an ulcerative and erosive right midfacial mass involving much of her right upper lip and extending from the vermillion border, superiorly into the right nasal vault. Infranasal examination revealed posterior extension well beyond the cartilaginous septum with significant erosion of the septum itself. No involvement of hard or soft palate intraorally. No palpable lymphadenopathy.

Diagnostic Workup and Results:

• Labs: Mild leukocytosis WBC = 11, Creatinine = 0.6, Urinalysis - negative for blood and protein, Utex – negative, Rheumatoid factor - negative, ANA - mildly positive at 8, C-ANCA - mild elevation at 1.80, RPR - non-reactive.

Case Presentation – Continued

Clinical Course

• Initially treated with multiple rounds of anti-biotics without improvement.
• Rheumatological consultation – Wegener’s Granulomatosis thought to be unlikely despite mildly positive C-ANCA due to atypical appearance of lesion, no granulomas or vasculitis on biopsy specimens, no systemic symptoms, ANCA atypical for Wegener’s due to perinuclear staining pattern.
• Infectious Disease consultation – felt unlikely to be of infectious etiology with no exposure risk factors and negative microbiological studies.
• Multidisciplinary tumor board discussion – felt malignancy ruled out.
• After 6 months of persistence of lesion without diagnosis patient was started on 60 mg oral prednisone daily for empiric trial which resulted in prompt and near-complete resolution of her ulcerative lesion within 1 month. Prednisone slowly tapered off after 2 months of treatment and patient without any evidence of recurrent lesion 2 months after discontinuing steroid therapy.
• Referred for facial plastics consultation due to loss of nasal tip support from persistent, but stable anterior septal destruction.

Discussion

Differential Diagnosis, Testing, and Treatment Approach

• Autoimmune/Vasculitis (Wegener’s, Lupus, Churg-Strauss) + C-ANCA, + ANA, +P-ANCA, urination for renal disease, vasculitis or granulomas on histology. Treat with immunomodulators - corticosteroids, methotrexate, cytotoxan.
• Malignancy (Squamous cell, basal cell, adenoid cystic carcinoma, NK/T cell lymphoma) - Diagnosis based on tissue biopsy – histopathology of fixed specimens for cutaneous malignancy and immunohistochemistry on fresh specimen for lymphoma. Treat with surgery for cutaneous malignancy and chemotherapy for lymphoma.
• Infectious (Syphilis, TB, Leprosy, Aspergillosis, Zygomycosis, Leishmaniasis) – Fungal, AFB, and culture specimens crucial. RPR testing. Immunocompromised, clinical history of exposure or pandemic areas. Treat with appropriate antimicrobial.
• Traumatic (Cocaine induced necrosis, Giant cell granuloma) – History of cocaine abuse, urine tox screen, C-ANCA often positive in cocaine users. Giant cells on histological examination. Cessation of cocaine and reconstructive repair, surgical excision for giant cell lesions.
• Unknown Cause (Sarcoidosis, Idiopathic midline destructive disease (IMMD)) Elevated serum ACE levels and non-caseating granulomas on histopathology for sarcoidosis. Diagnosis of exclusion for IMMD. Treat sarcoidosis with corticosteroids, cytotoxan, infliximab. IMMD treated with local radiation, cytotoxan and corticosteroids.

Conclusions

• The differential diagnosis for destructive lesions of the midline face is wide and includes numerous benign and malignant processes.
• A multidisciplinary approach and thorough tissue biopsy with emphasis on both fresh and permanent specimens is crucial to diagnosis, yet in some instances no etiology will be found.
• The otorhinolaryngologist should be aware of the differential diagnosis of midline destructive lesions as a disease entity to ensure timely diagnostic efforts and treatment initiation.

References